Cardiac function in the Infant with CDH

Neil Patel
Royal Hospital for Children
Glasgow, UK

neil.patel@ggc.scot.nhs.uk

PEOPLE MAKE GLASGOW

NHS Greater Glasgow and Clyde

Neonatal management?

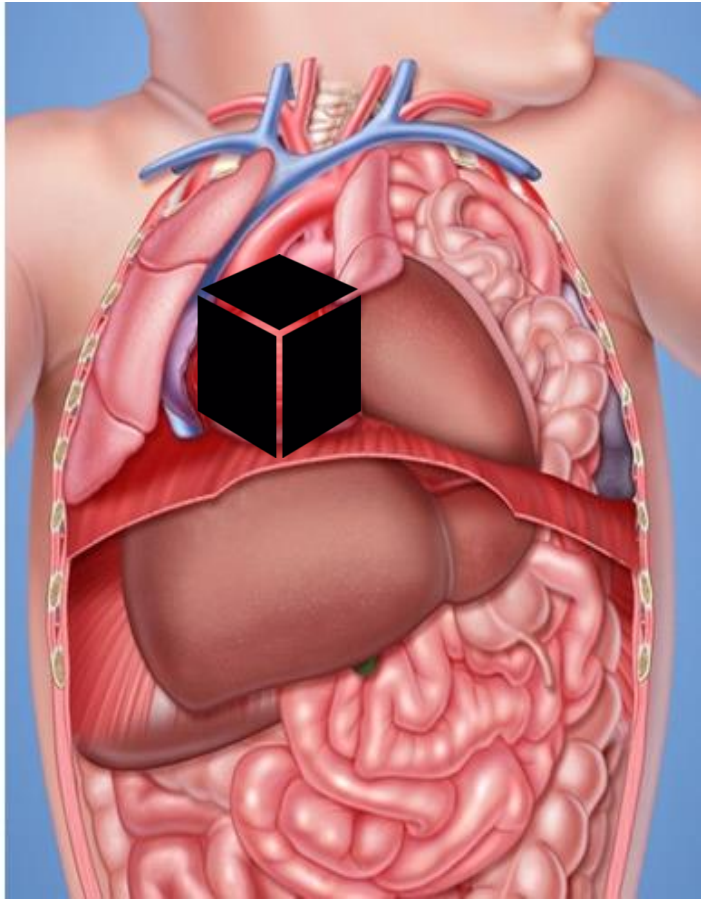
Timing of surgery?

Delivery room management?

ECLS /ECMO?

Prenatal prediction?

Long term outcome?



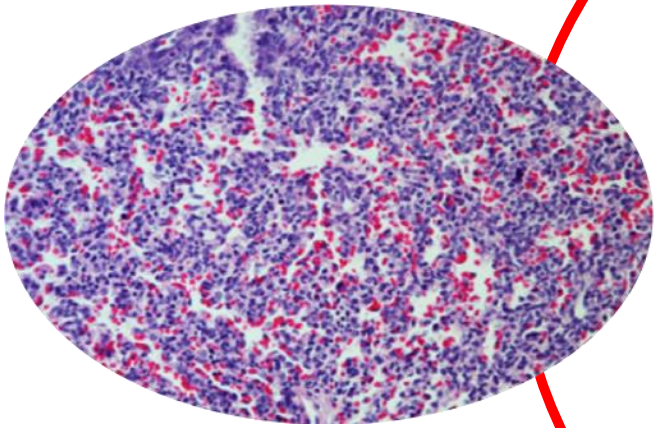
Cardiac function in CDH

Pathophysiology

Assessment & Management

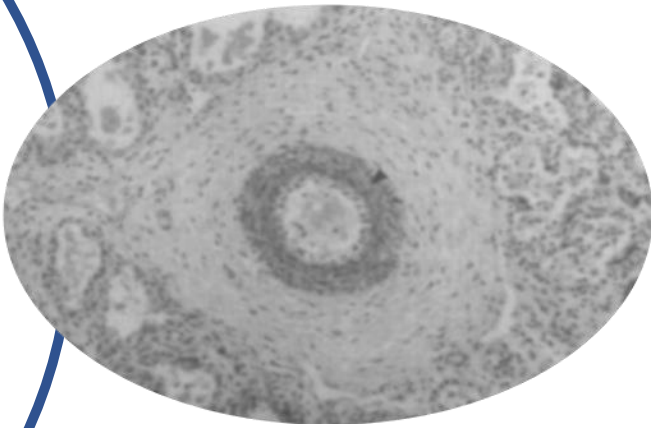
Future directions

Pulmonary hypoplasia



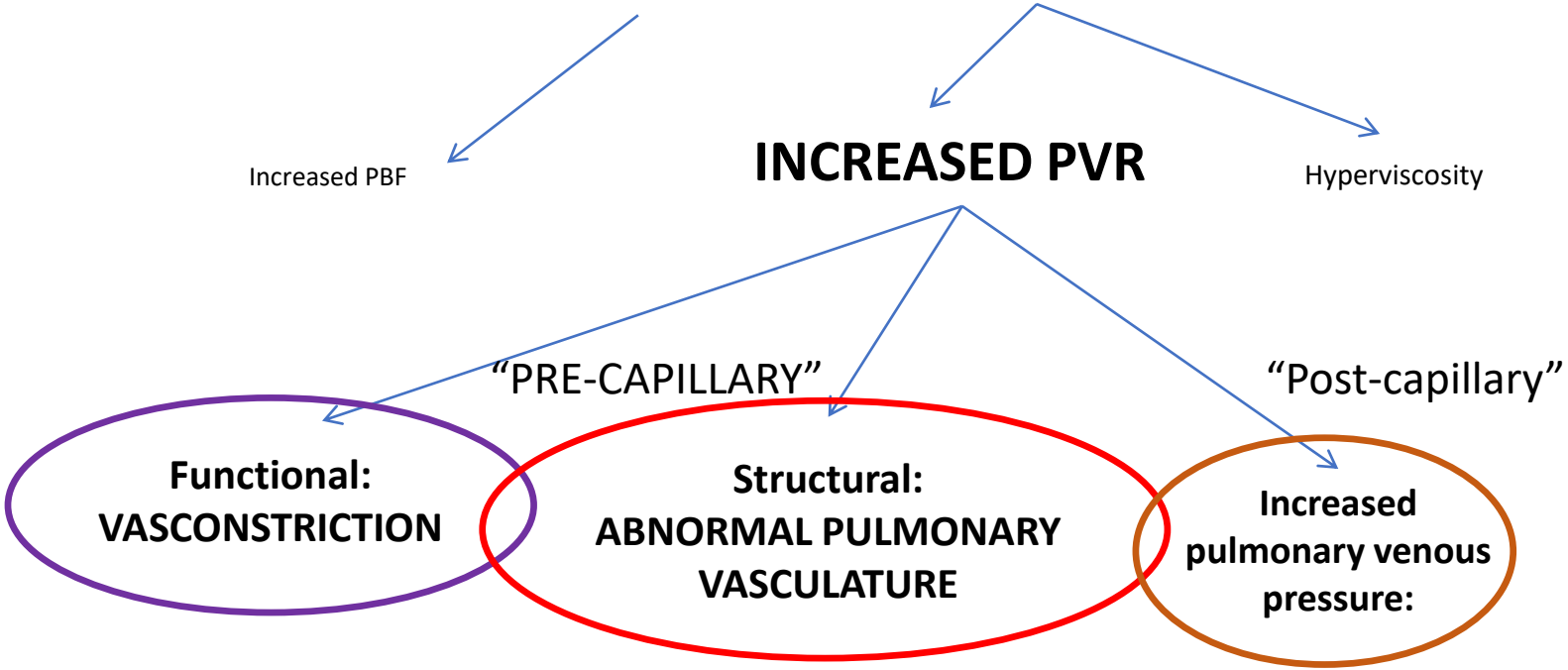
Schmidt et al, 2012

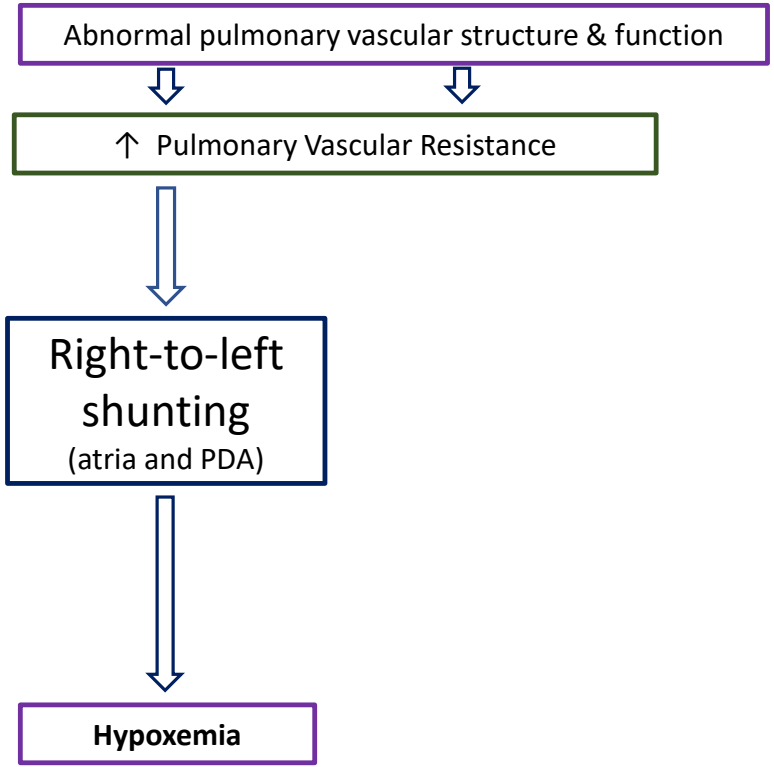
Pulmonary hypertension

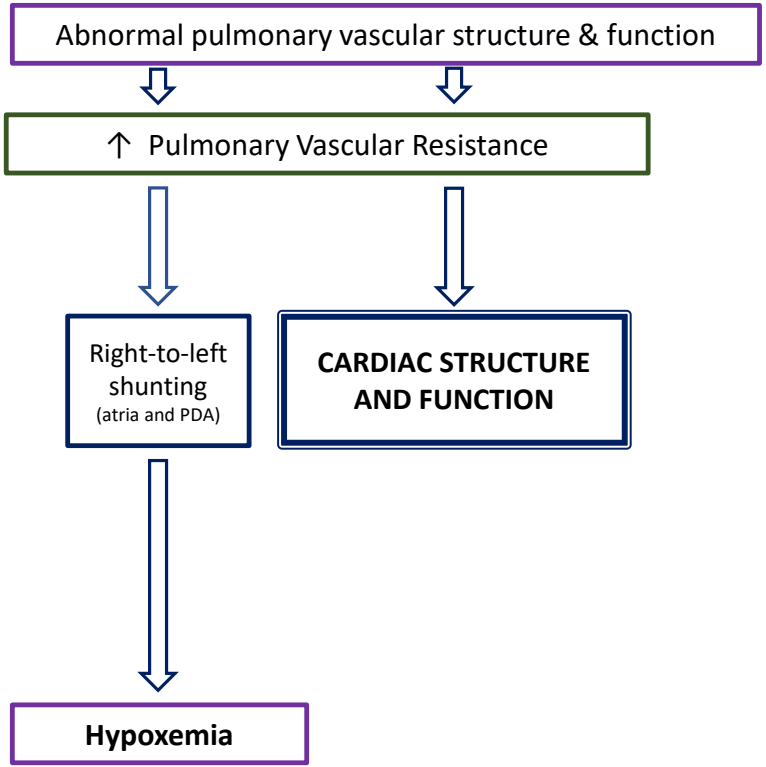


Yamataka and Puri, 1997

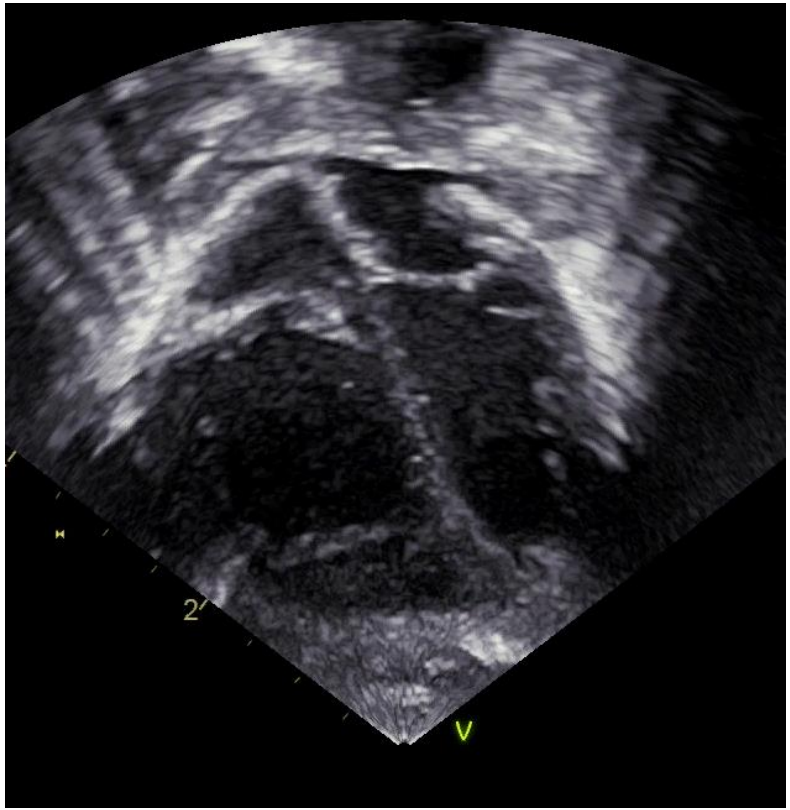
↑PAP = PBFlow x PVRESISTANCE

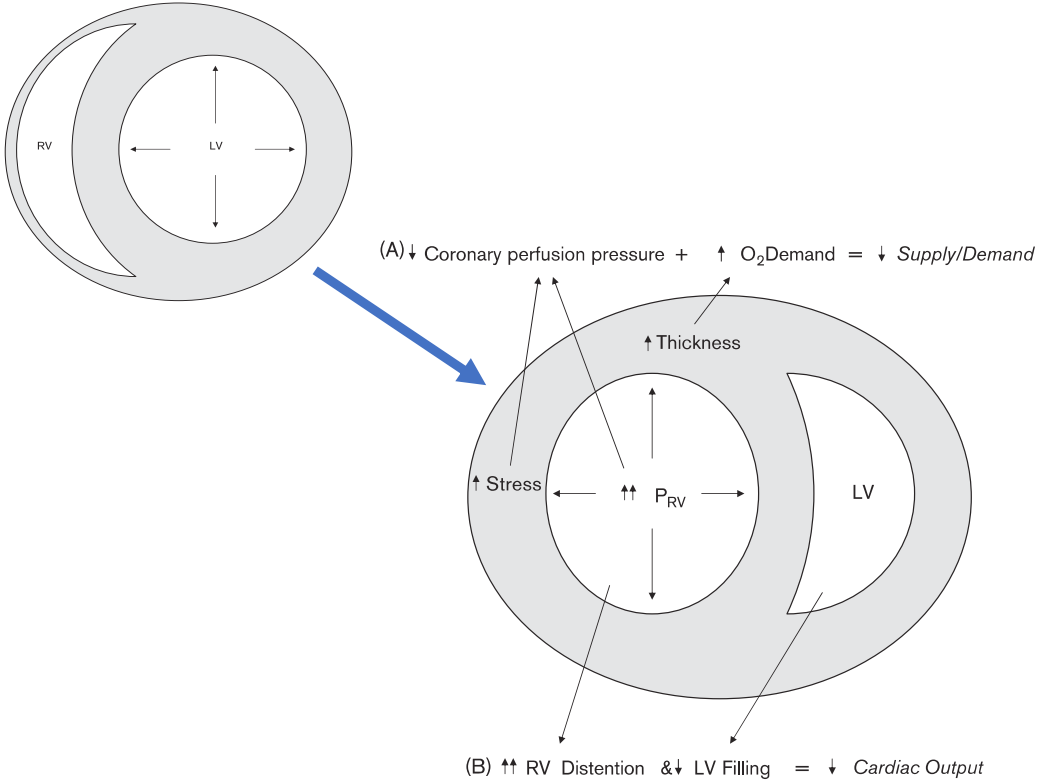



Abnormal pulmonary vascular structure & function
↑ Pulmonary Vascular Resistance
Right-to-left shunting
(atria and PDA)
Hypoxemia

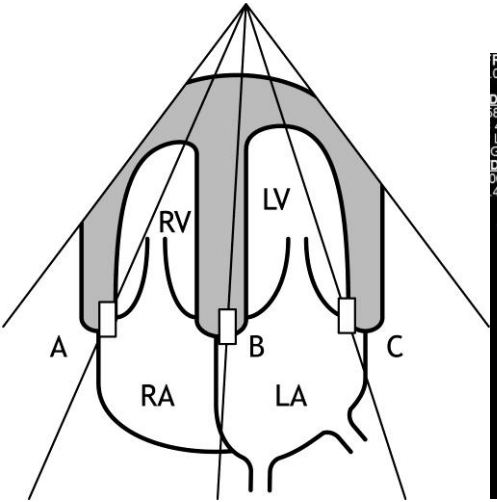

Abnormal pulmonary vascular structure & function
↑ Pulmonary Vascular Resistance
Right-to-left shunting
(atria and PDA)
CARDIAC STRUCTURE AND FUNCTION
Hypoxemia

RV dilatation and hypertrophy in pulmonary hypertension

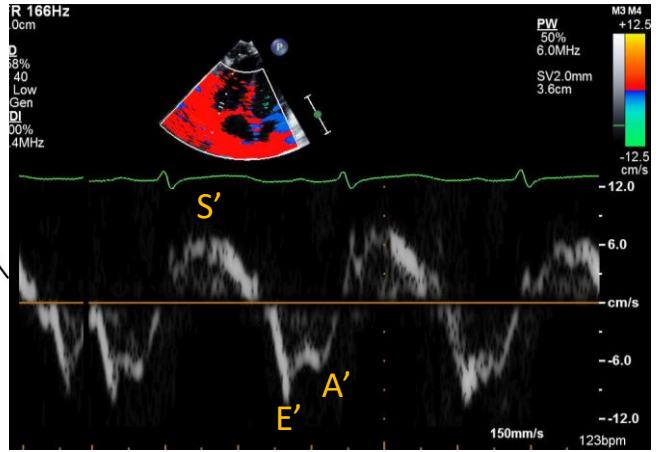




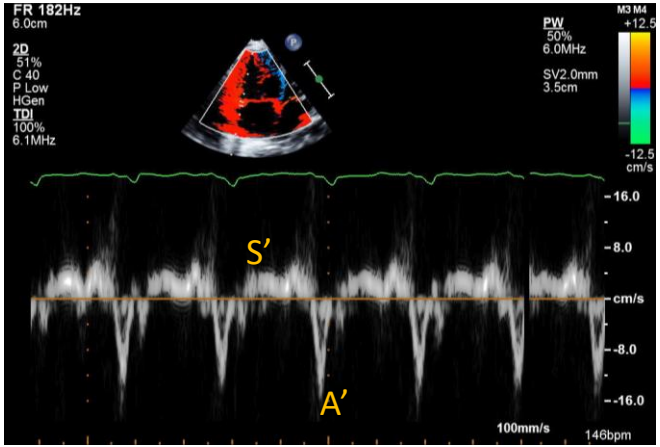
Tissue Doppler Imaging of RV in CDH



Control

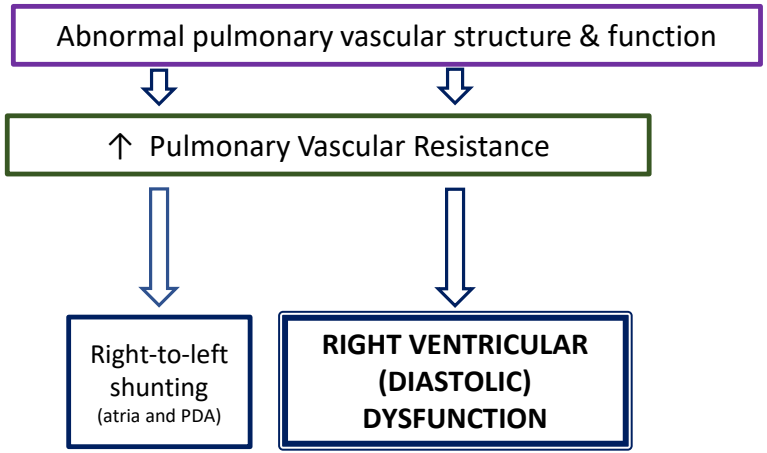


CDH

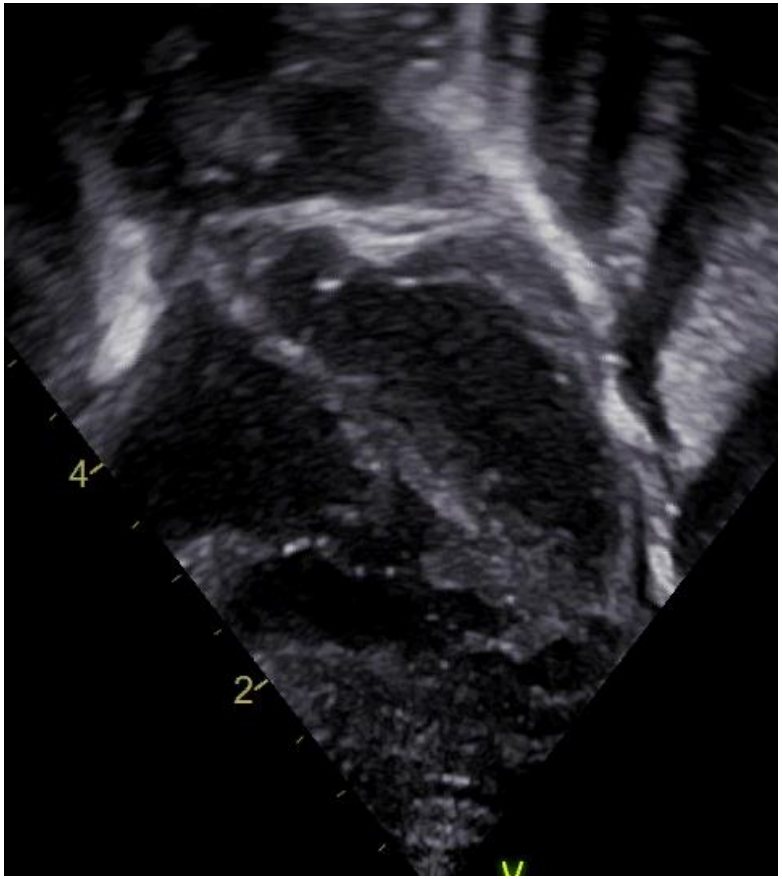


- Reduced systolic velocities (S')
- **Loss of diastolic (e') velocity**

Patel et al, Neonatology 2009


Abnormal pulmonary vascular structure & function
↑ Pulmonary Vascular Resistance
Right-to-left shunting
(atria and PDA)
RIGHT VENTRICULAR (DIASTOLIC) DYSFUNCTION

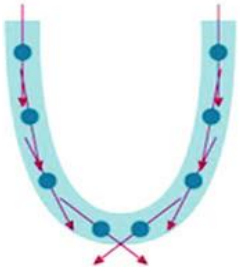
CDH Day 1: FiO_2 1.0, hypotensive, acidotic



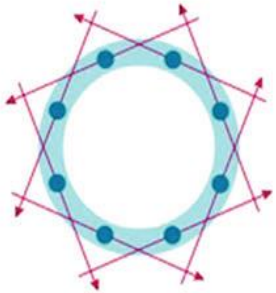
Speckle tracking echocardiography in CDH

NHS Greater Glasgow and Clyde

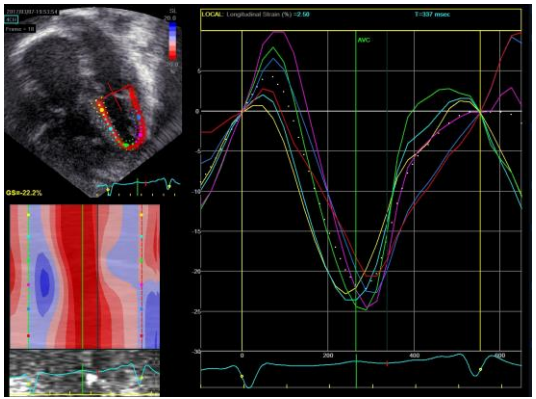
Longitudinal strain (LS)

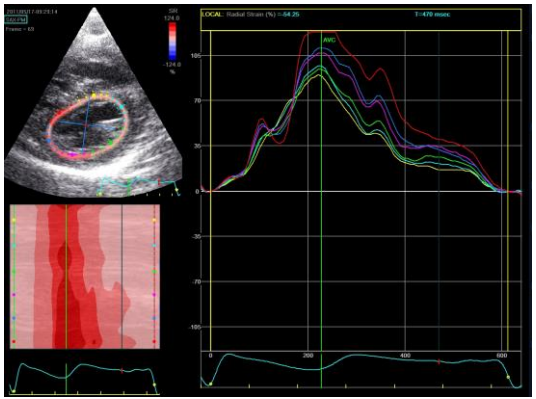


Circumferential strain (CS)

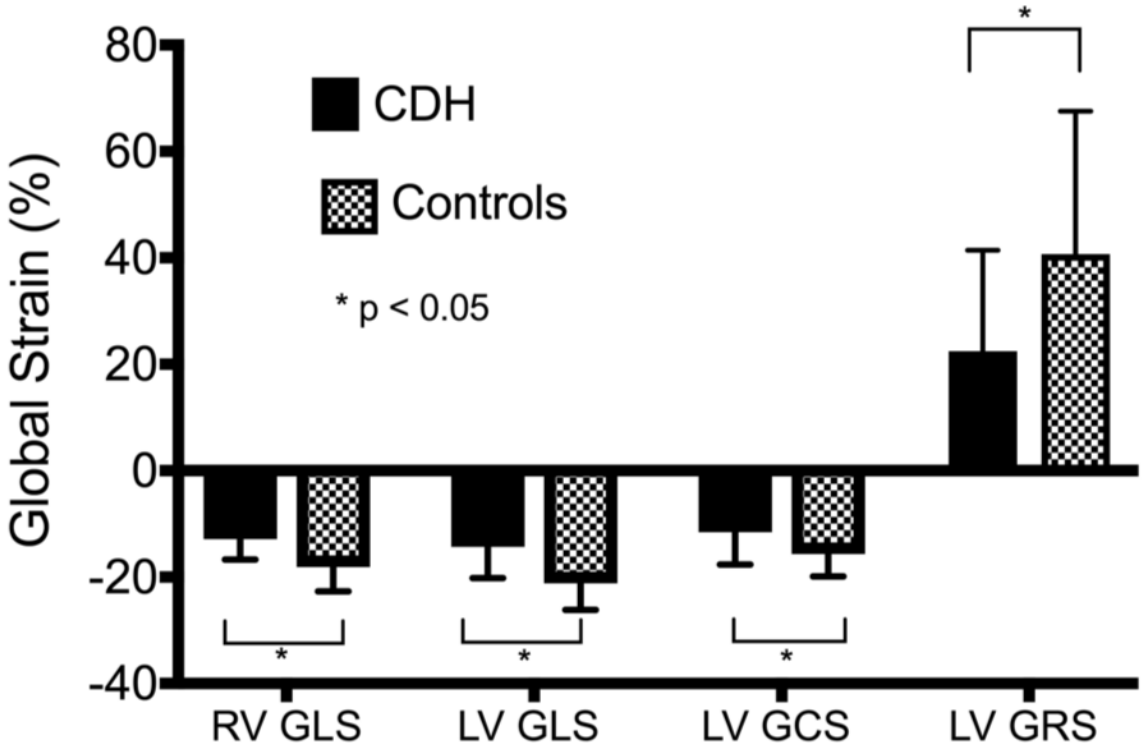


Radial strain(RS)

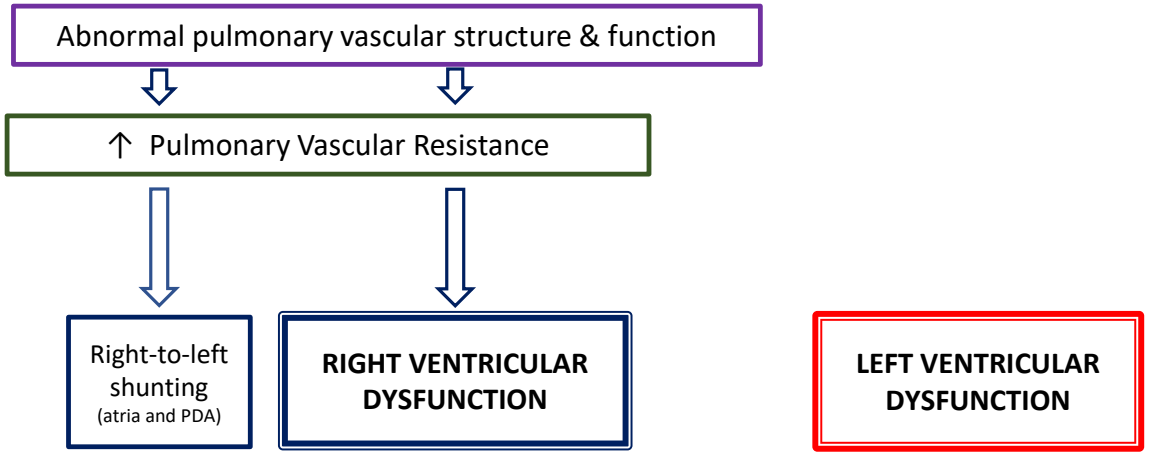


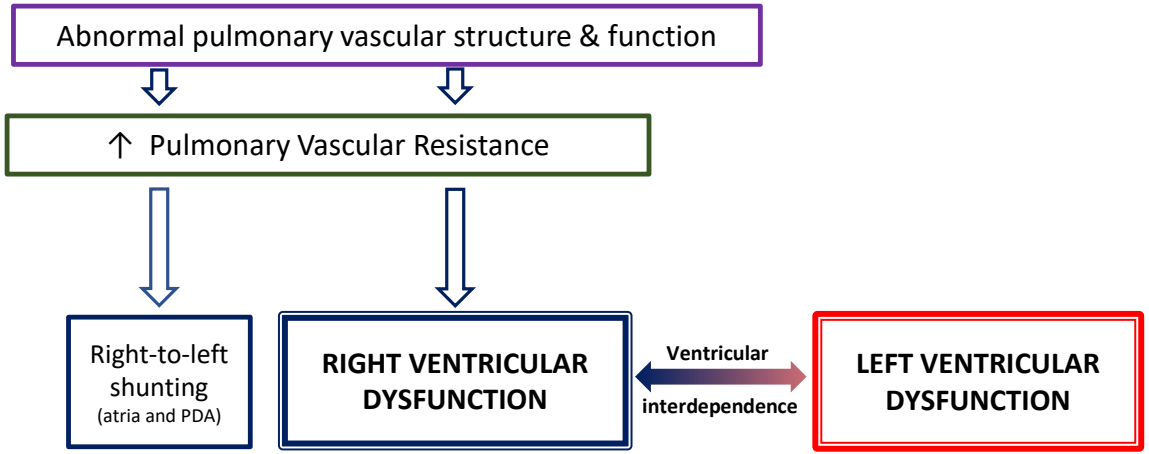


Ventricular strain in CDH (first 48h of life)



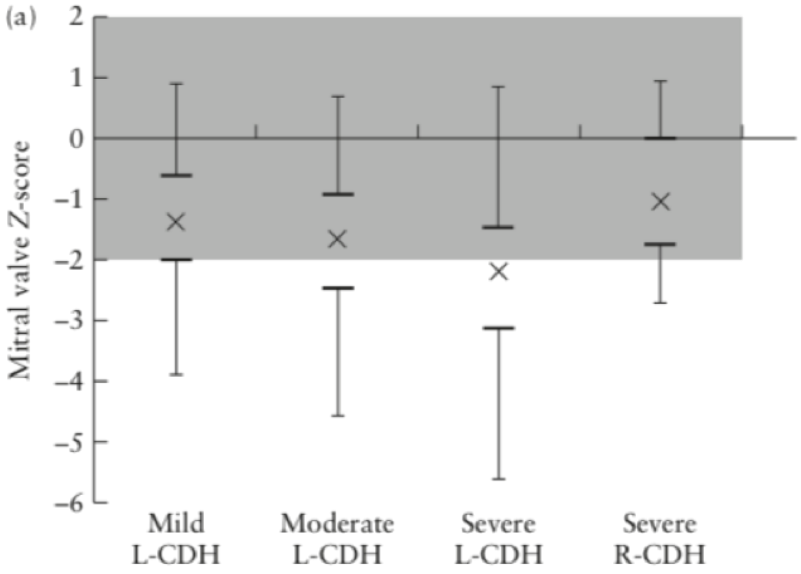
Patel et al, J Peds 2018

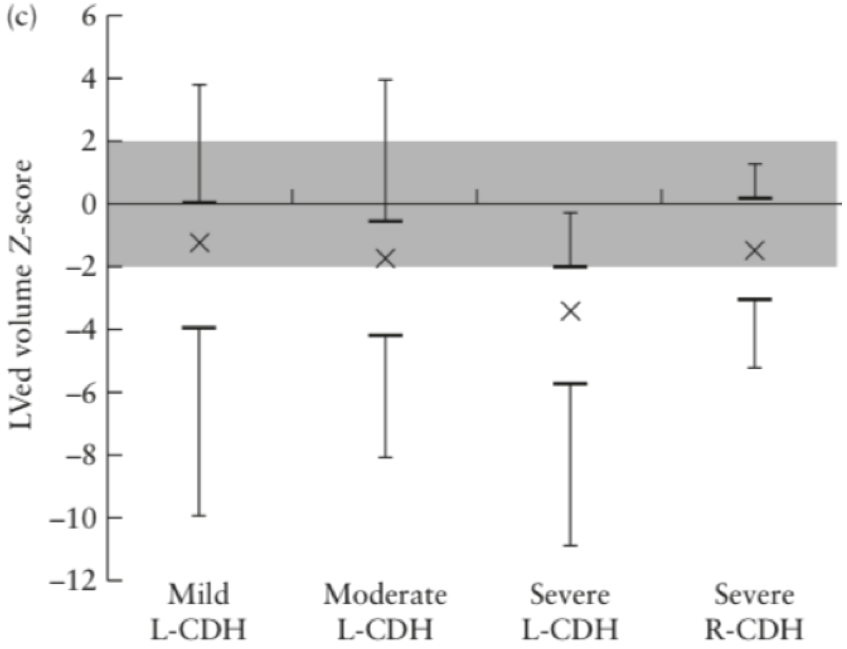

Abnormal pulmonary vascular structure & function
↑ Pulmonary Vascular Resistance
Right-to-left shunting
(atria and PDA)
RIGHT VENTRICULAR DYSFUNCTION
LEFT VENTRICULAR DYSFUNCTION


Abnormal pulmonary vascular structure & function
↑ Pulmonary Vascular Resistance
Right-to-left shunting
(atria and PDA)
RIGHT VENTRICULAR DYSFUNCTION
Ventricular interdependence
LEFT VENTRICULAR DYSFUNCTION

Severe left diaphragmatic hernia limits size of fetal left heart more than does right diaphragmatic hernia

F. A. BYRNE*, R. L. KELLER†, J. MEADOWS*, D. MINIATI‡§, M. M. BROOK*, N. H. SILVERMAN* and A. J. MOON-GRADY*§





Ultrasound Obstet Gynecol 2015; **46**: 688–694

Possible mechanisms of fetal LV hypoplasia

- Reduced pulmonary blood flow
- Altered ductus venosus streaming
- Mechanical compression

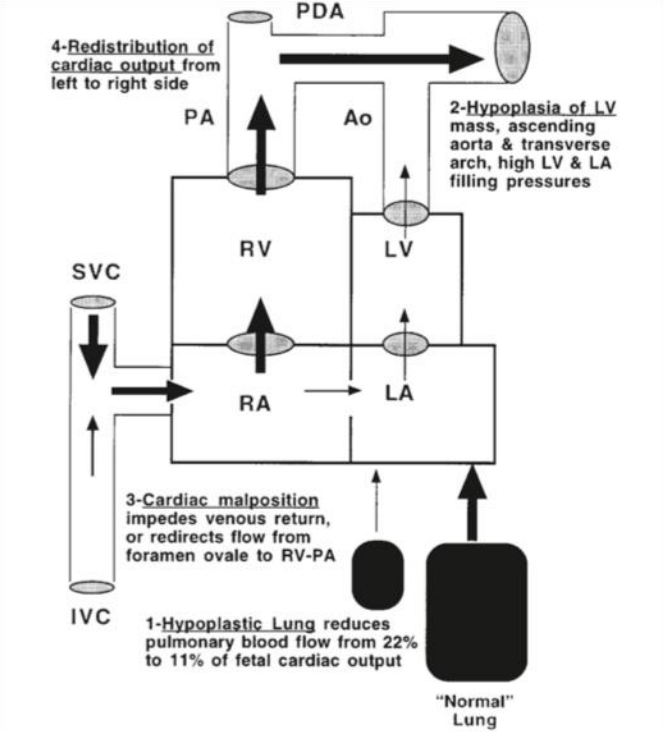
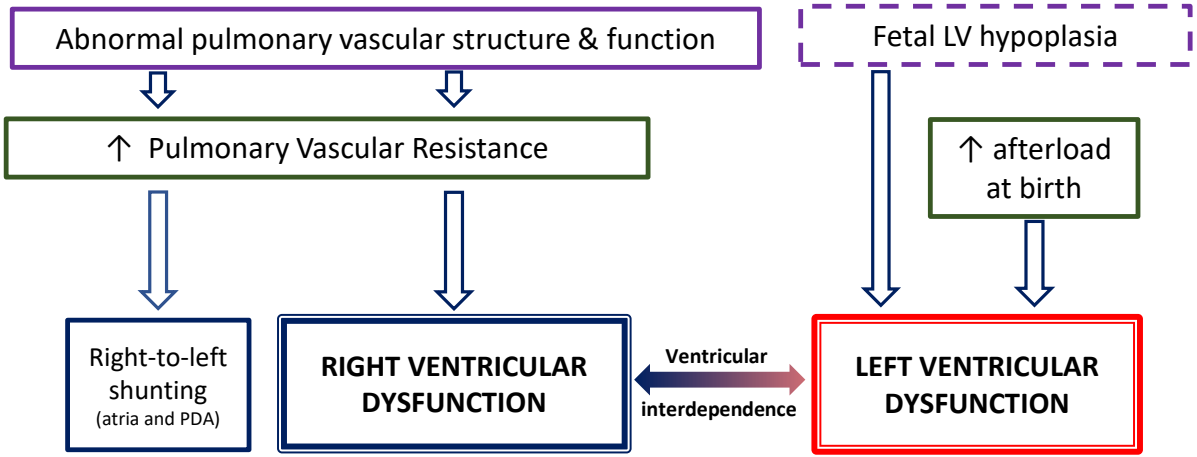
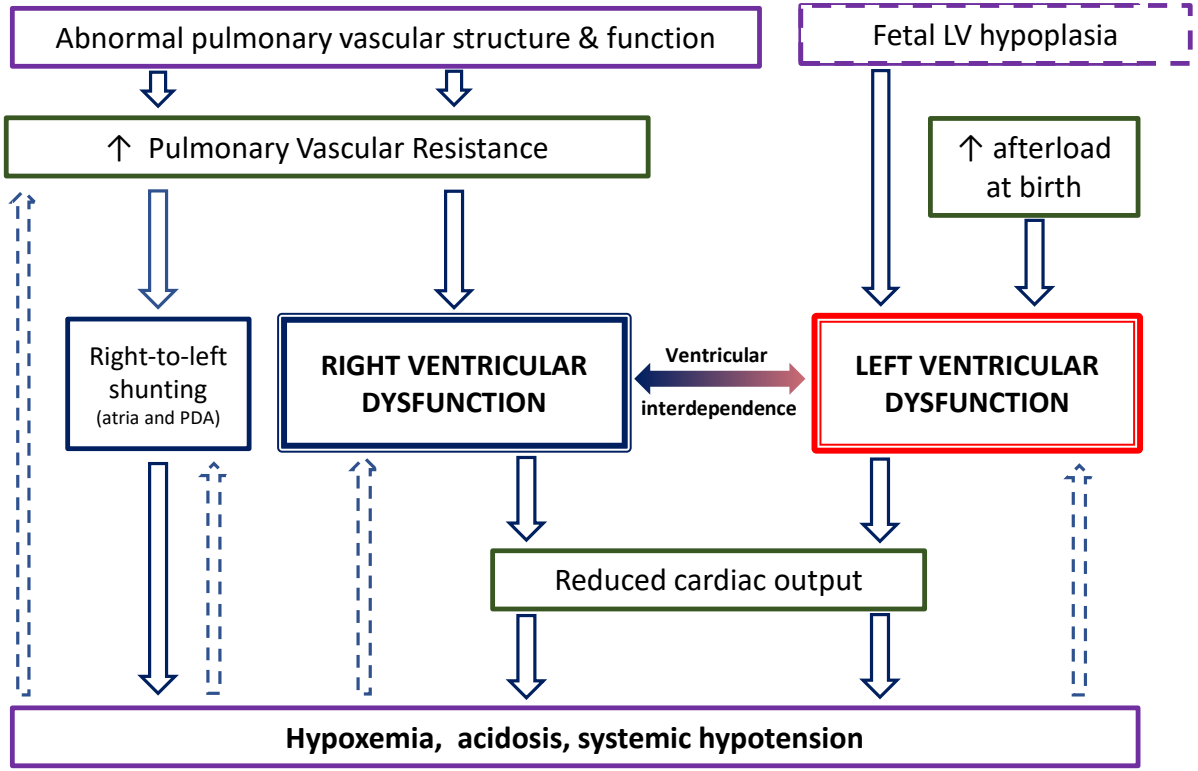


Fig. 2. Diagram summarizing four mechanisms for cardiovascular compromise during fetal life with severe CDH. (Adapted with permission from WB Saunders. Katz AL, Wiswell TE, Baumgart S. Clin Perinatol 1998.)[17]

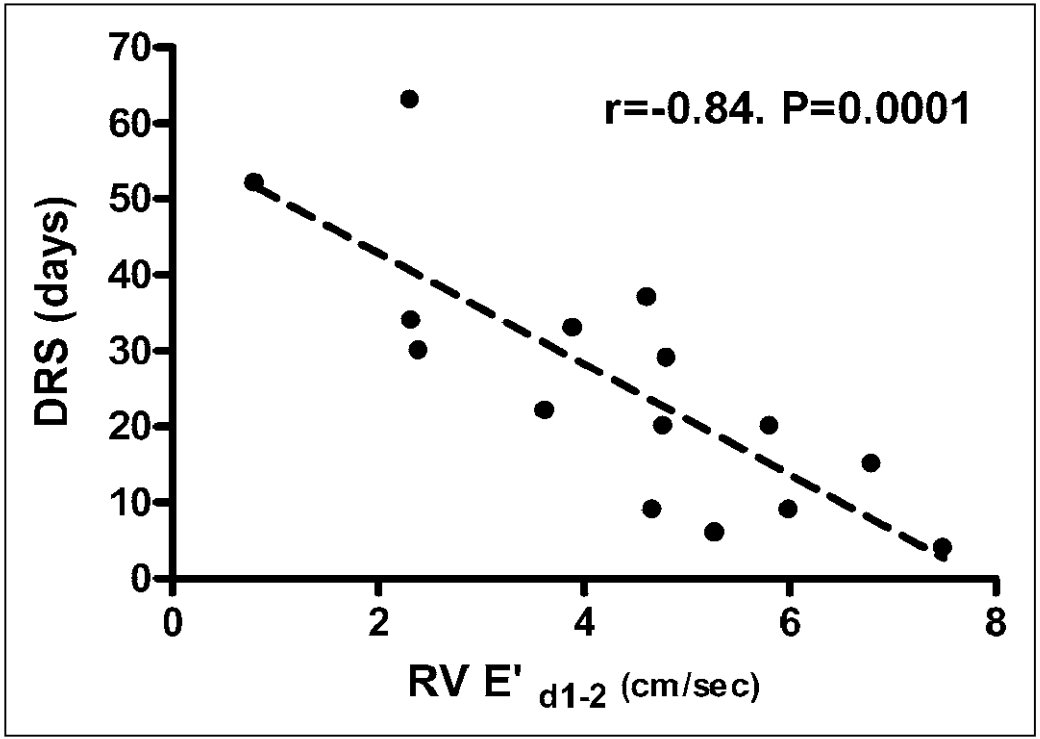

Abnormal pulmonary vascular structure & function
Fetal LV hypoplasia
↑ Pulmonary Vascular Resistance
↑ afterload at birth
Right-to-left shunting (atria and PDA)
RIGHT VENTRICULAR DYSFUNCTION
Ventricular interdependence
LEFT VENTRICULAR DYSFUNCTION



Patel et al, Sem Perinatol 2019

Right Ventricular Diastolic Function Measured by Tissue Doppler Imaging Predicts Early Outcome in Congenital Diaphragmatic Hernia

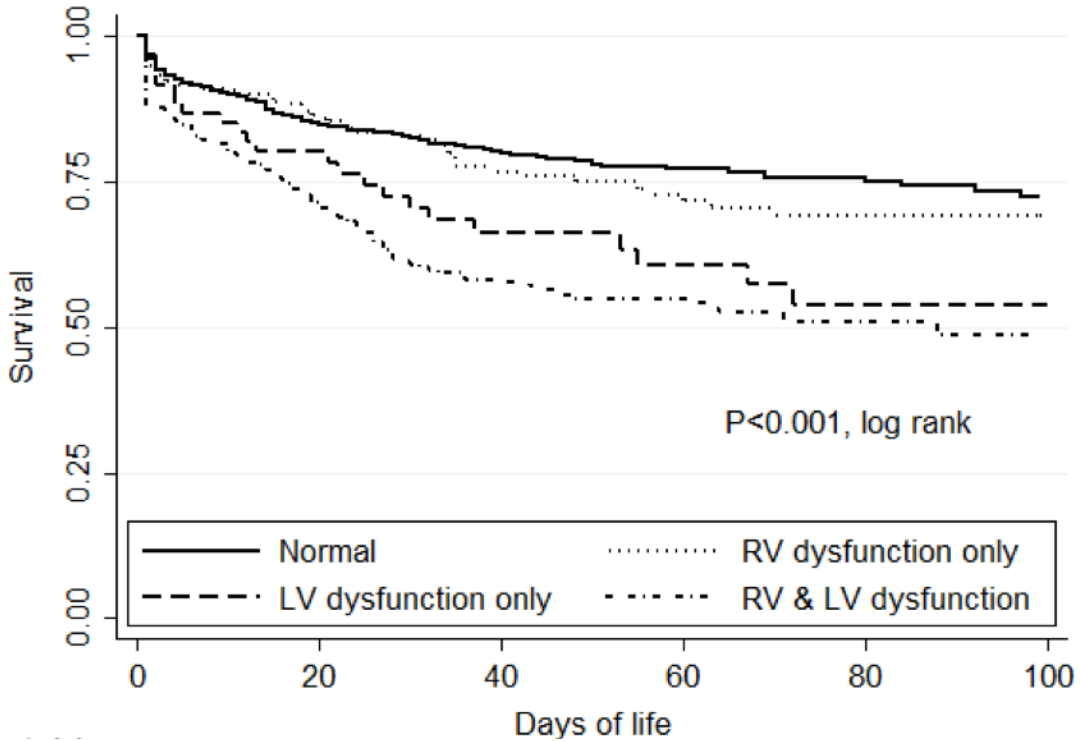
Florian Moenkemeyer, MD; Neil Patel, MD



Ped Crit Care Med 2013

Ventricular Dysfunction is a Critical Determinant of Mortality in Congenital Diaphragmatic Hernia

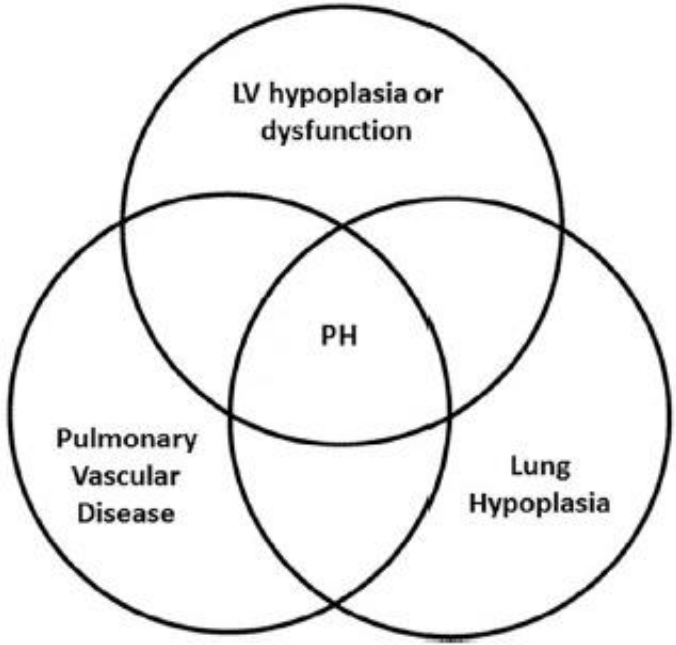
Neil Patel , Pamela A Lally , Florian Kipfmueller , Anna Claudia Massolo , Matias Luco , Krisa P Van Meurs , Kevin P Lally , Matthew T Harting , and , for the Congenital Diaphragmatic Hernia Study Group



AJRCCM 2019

The Left Ventricle in Congenital Diaphragmatic Hernia: Implications for the Management of Pulmonary Hypertension

- Pulmonary Venous Hypertension
- Decreased cardiac output
- Pulmonary edema, worsened with PH drug therapy

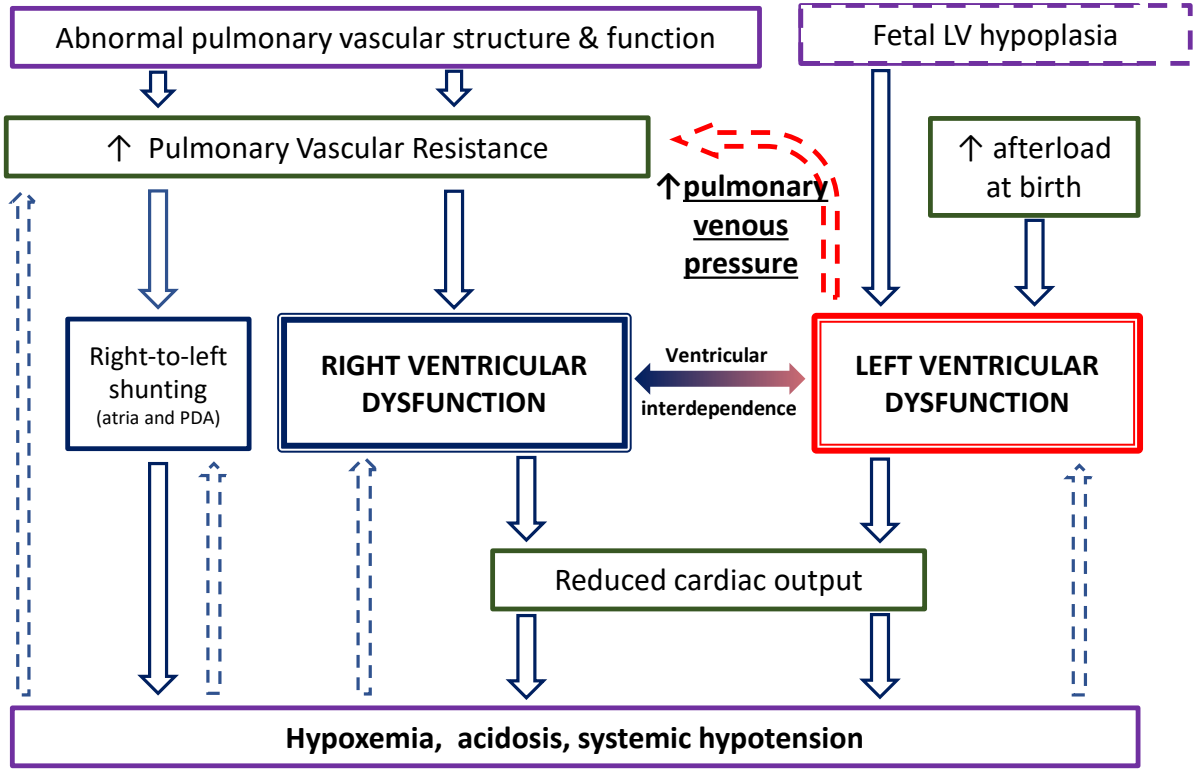


- High tone and abnormal reactivity;
- structural remodeling
- decreased vessel growth

- Decreased phasic lung stretch and/or compression of lung,
- decreased surface area for gas exchange,
- poor lung recruitment and lung injury from mechanical ventilation

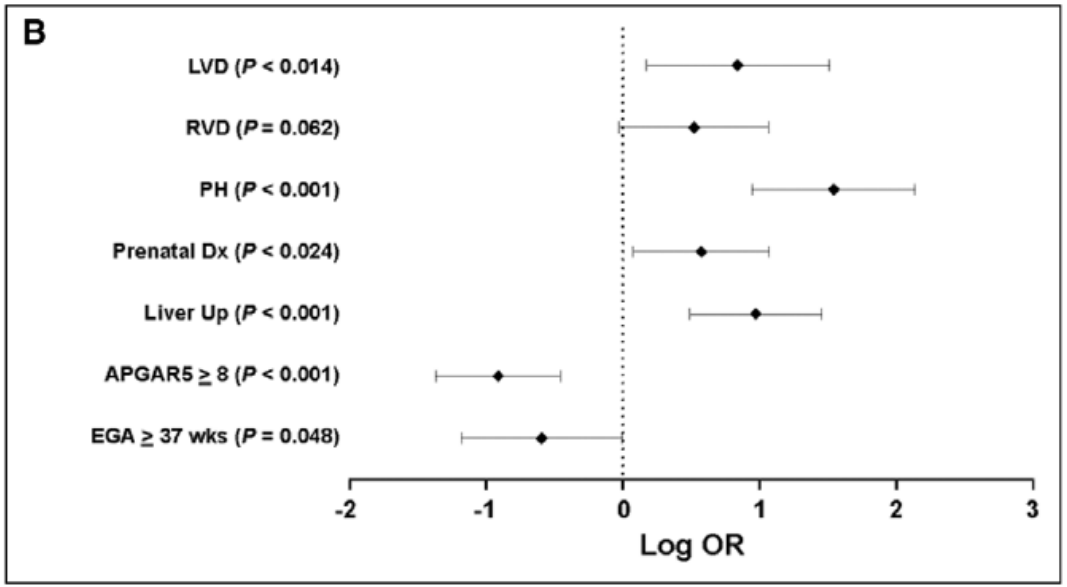
Kinsella et al, J Peds, 2

LV dysfunction may cause ↑PVR



Early Left Ventricular Dysfunction and Severe Pulmonary Hypertension Predict Adverse Outcomes in "Low-Risk" Congenital Diaphragmatic Hernia

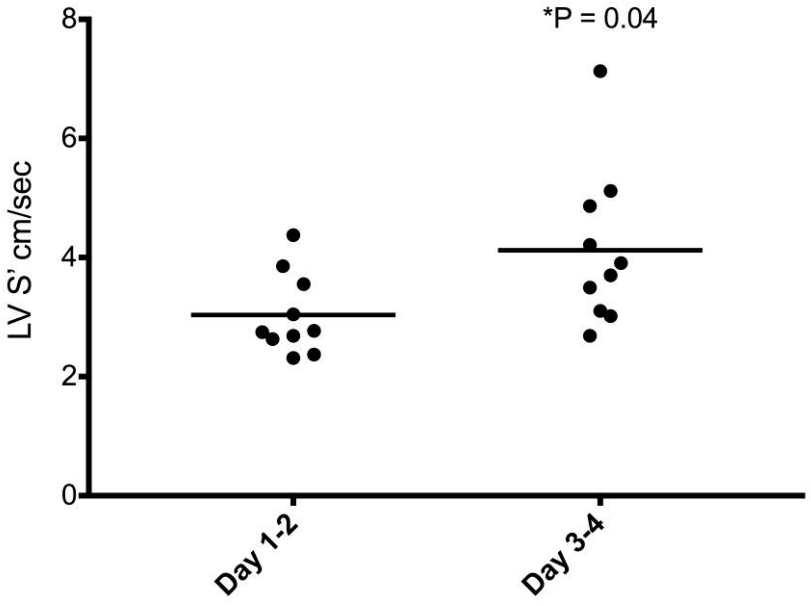
Duy T. Dao, MD, MPH[1,2]; Neil Patel, MB ChB, MD[3]; Matthew T. Harting, MD, MS[4]; Kevin P. Lally, MD, MS[4]; Pamela A. Lally, MD[4]; Terry L. Buchmiller, MD[1]



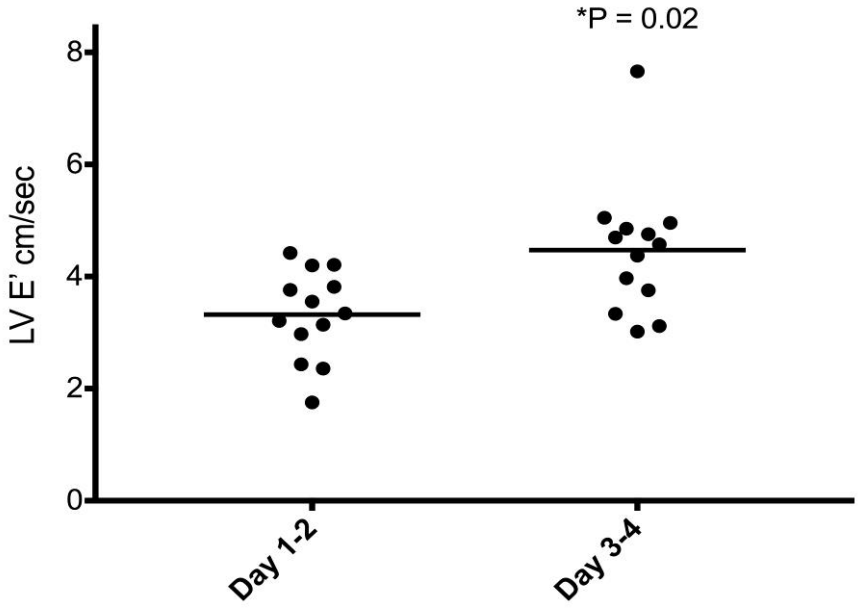
PCCM 2020

LV function improves in first days of life

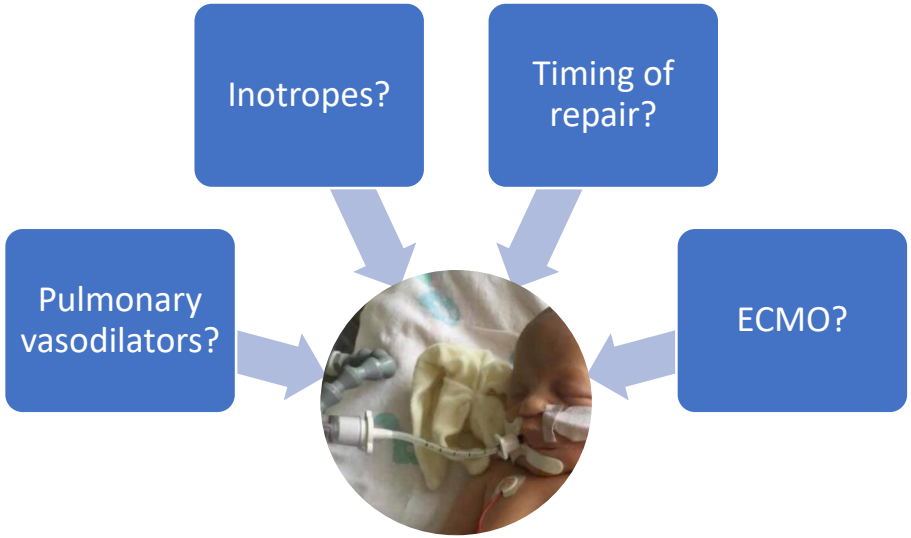
LV systolic function (TDI LV S')

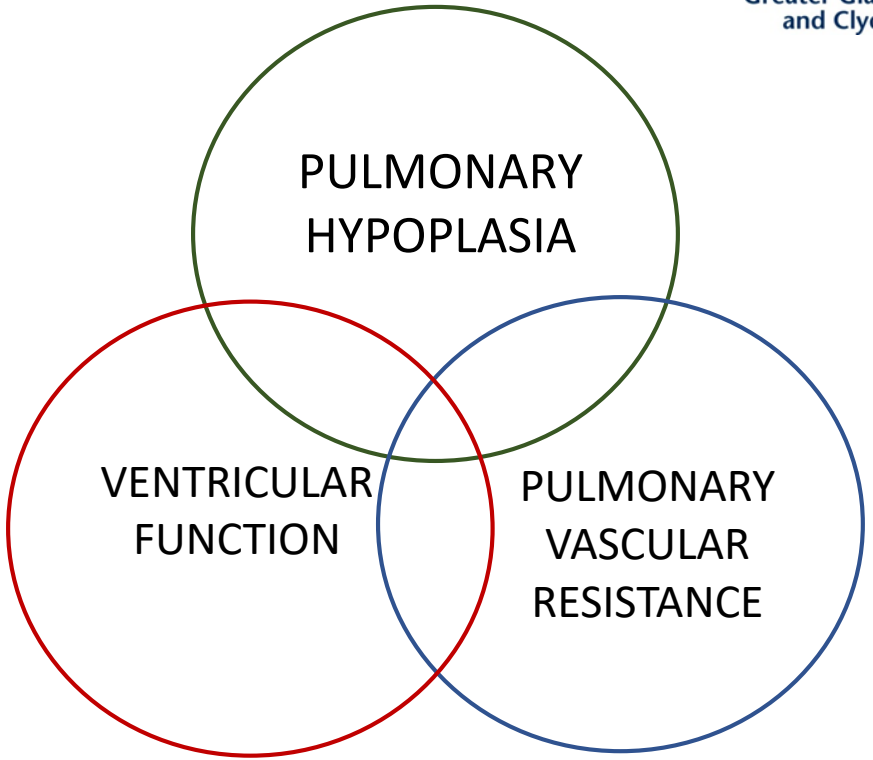


LV diastolic function (TDI LV E')



N. Patel, F. Kipfmueller / Seminars in Pediatric Surgery 26 (2017)


Inotropes?
Timing of repair?
Pulmonary vasodilators?
ECMO?


PULMONARY HYPOPLASIA
VENTRICULAR FUNCTION
PULMONARY VASCULAR RESISTANCE

Continuous multi-modal assessment:

PULMONARY HYPOPLASIA	PULMONARY HYPERTENSION		CARDIAC FUNCTION RV and LV
	Oxygenation / oxygen delivery	**PVR / PAP**	
Prenatal lung volumes	SaO_2	Pre-post ductal saturations	Systemic Blood Pressure: pulse pressure
(Postnatal lung volume- ?CXR?)	PaO_2	**Echo assessment:** -Tricuspid regurgitation velocity -PDA shunting pattern -Time to peak velocity in pulmonary artery -Septal shape	**Echo assessment:** "Eyeballing" from 2d loops Quantitative measures: ➢ Tissue Doppler imaging ➢ Speckle tracking echocardiography
PCO_2 $ETCO_2$ Transcutaneous CO_2	Lactate Cardiac output NIRS / SVO2 aEEG		
Ventilator settings			

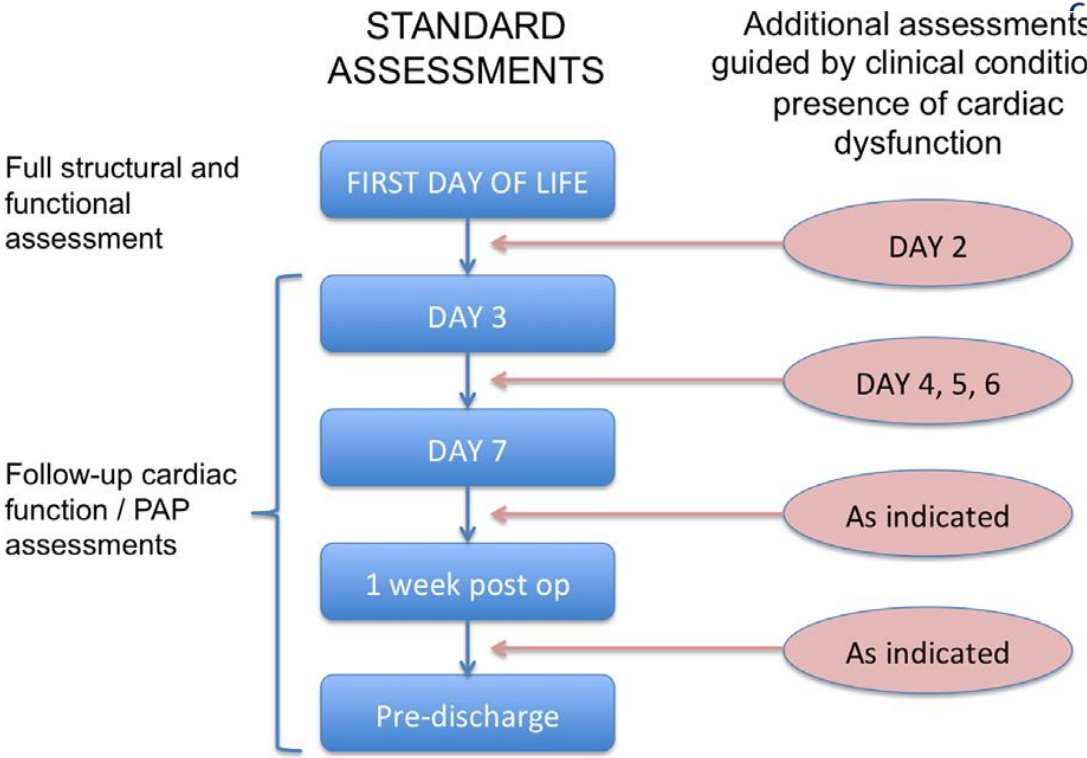
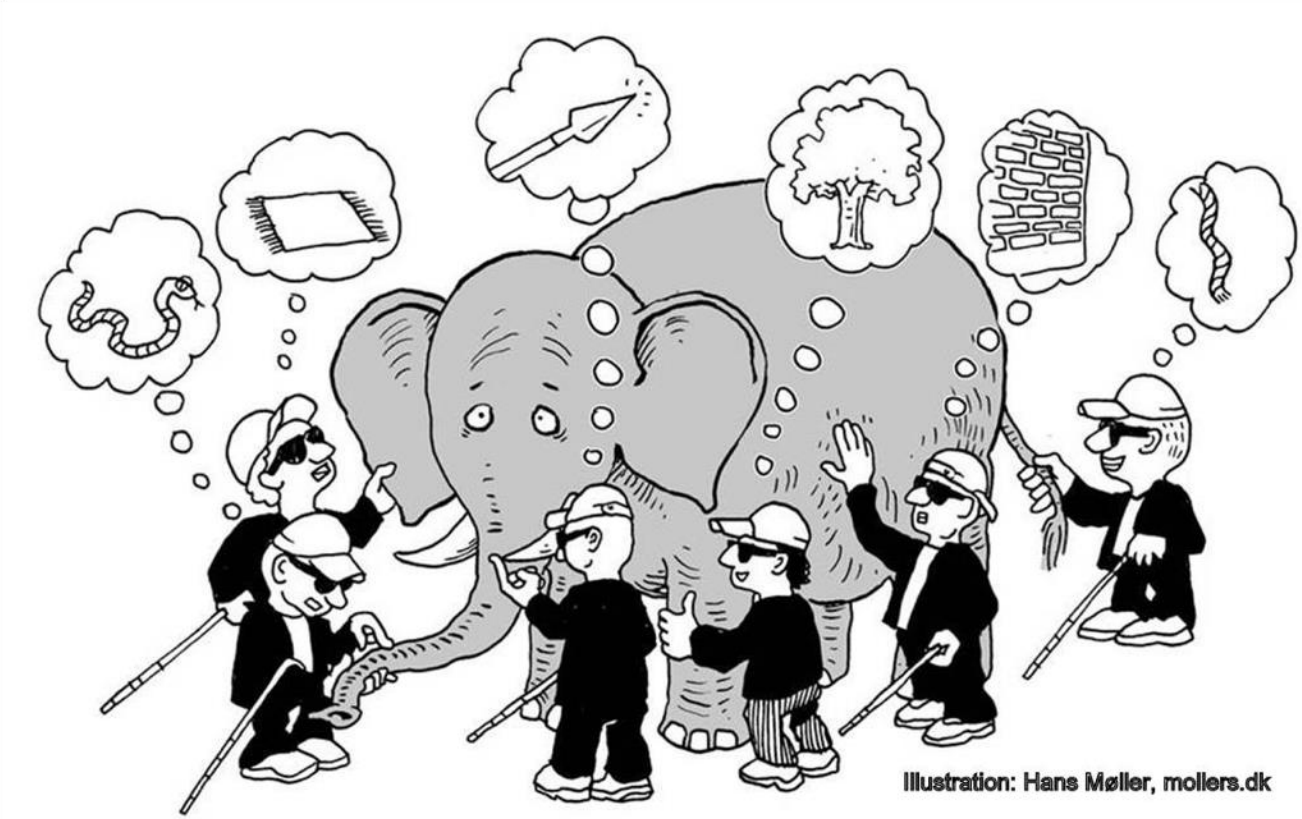


Fig. 3. Example protocol for timing of cardiac function assessment in CDH.

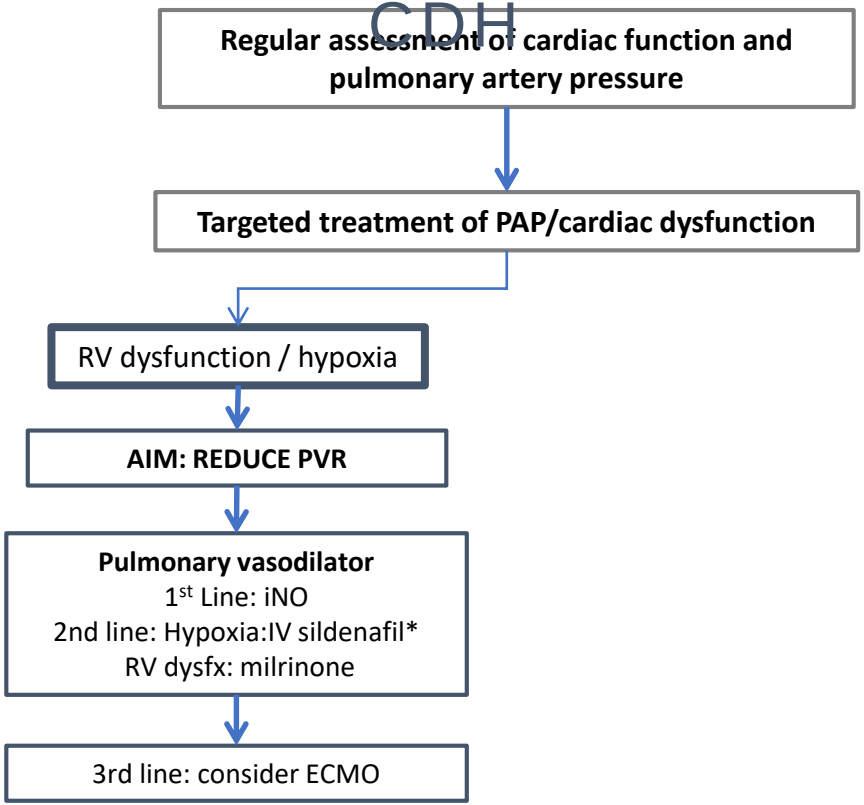
N. Patel, F. Kipfmueller / Seminars in Pediatric Surgery 26 (2017)

The Blind Men of Indostan and the Elephant in the Echo Lab

Lawrence G. Rudski, MDCM, FACC, FASE, and Jonathan Afilalo, MD, MSc, FRCPC, *Montreal, Quebec, Canada*



Targeted therapy of PH and cardiac function in CDH



Targeted therapy of PH and cardiac function in CDH

Regular assessment of cardiac function and pulmonary artery pressure

↓

Targeted treatment of PAP/cardiac dysfunction

↓

RV dysfunction / hypoxia

↓

AIM: REDUCE PVR

↓

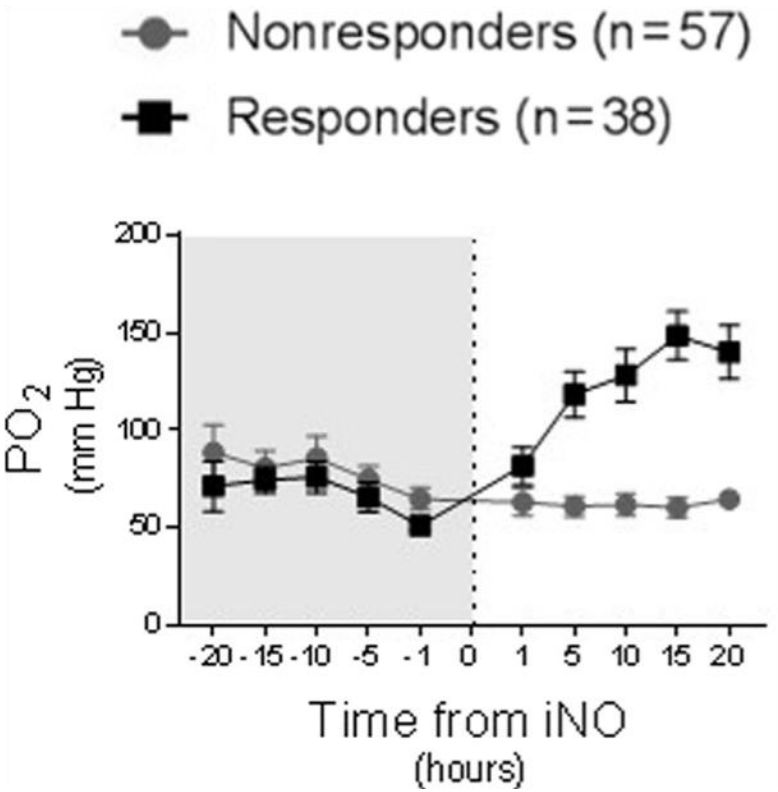
Pulmonary vasodilator
1st Line: iNO
2nd line: Hypoxia:IV sildenafil*
RV dysfx: milrinone

↓

3rd line: consider ECMO

PGE_1 to
maintain PDA if:
- RV dilatation/ dysfx
- PAP ≥ systemic BP
- constricting ductus arteriosus

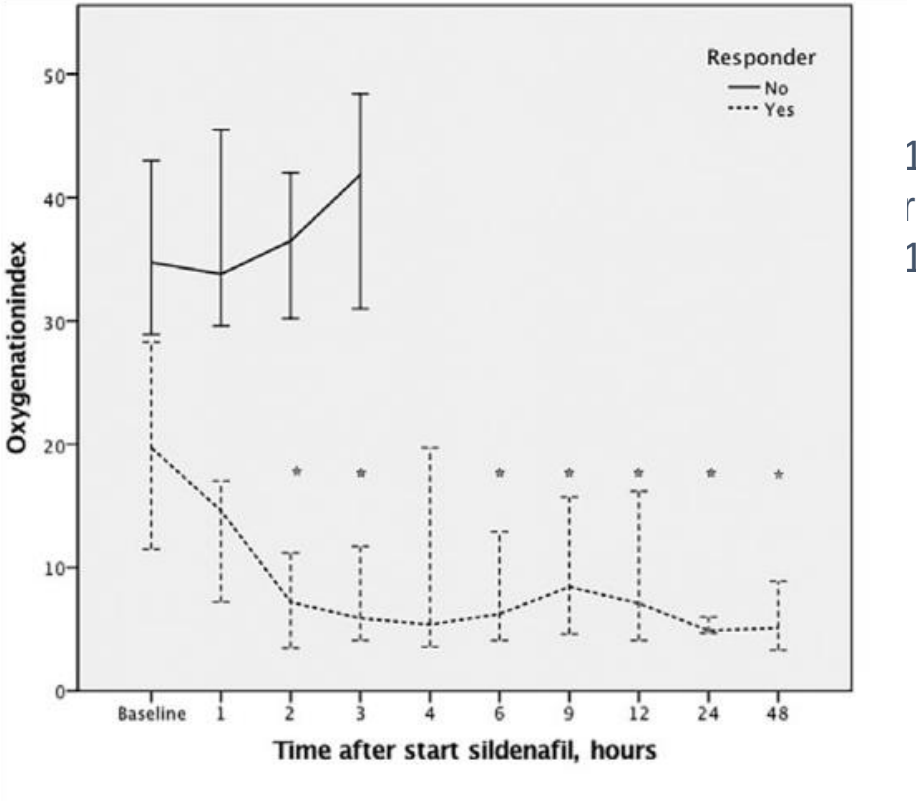
Inhaled Nitric Oxide Is Associated with Improved Oxygenation in a Subpopulation of Infants with Congenital Diaphragmatic Hernia and Pulmonary Hypertension



Lawrence et al, J Peds 2019

Continuous intravenous sildenafil as an early treatment in neonates with congenital diaphragmatic hernia

Florian Kipfmueller MD[1] | Lukas Schroeder MD[1] | Christoph Berg MD[2] | Katrin Heindel MD[1] | Peter Bartmann MD, PhD[1] | Andreas Mueller MD[1]



15 non-responders
11 responders

Ped Pulmonol 2018

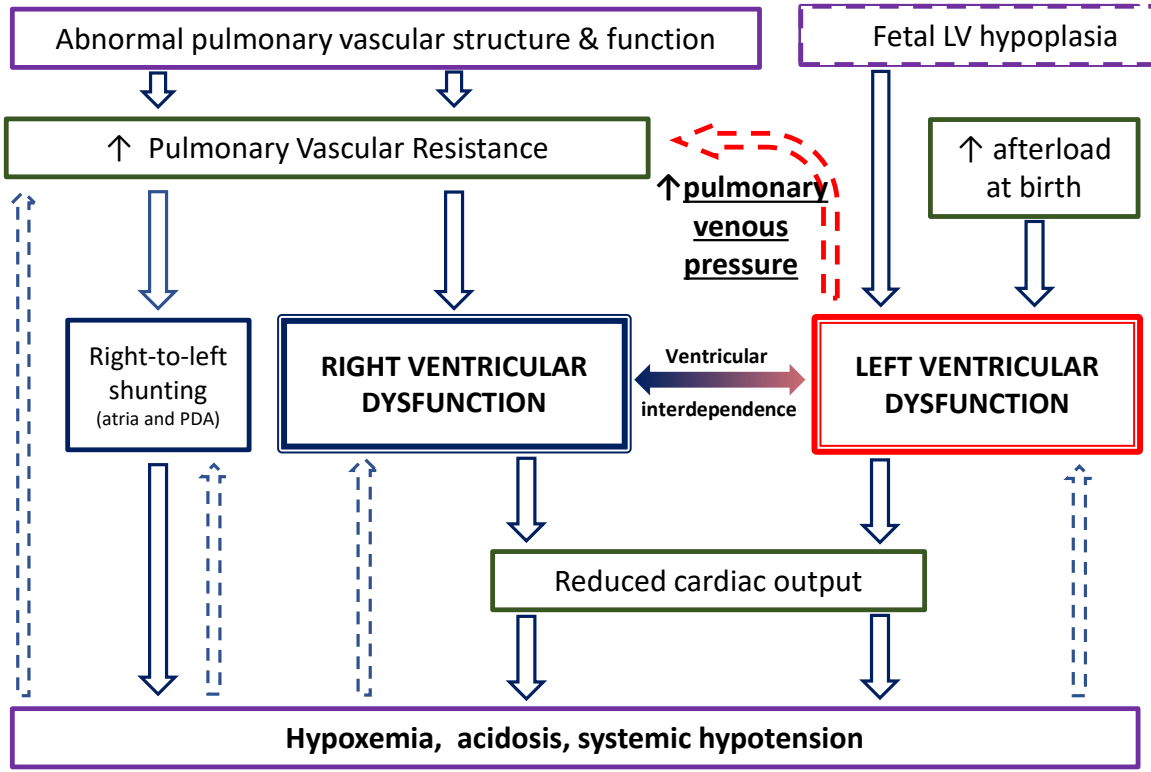
Inhaled Nitric Oxide Is Associated with Improved Oxygenation in a Subpopulation of Infants with Congenital Diaphragmatic Hernia and Pulmonary Hypertension

NHS Greater Glasgow and Clyde

Table I. Characteristics of initial responders and nonresponders to iNO therapy

Patient characteristics	Nonresponder to iNO therapy (n = 57)	Responder to iNO therapy (n = 38)	*P* value
Male sex (n, %)	28 (49%)	23 (61%)	.30
Gestational age (wk)	38.0 ± 0.2	38.4 ± 0.2	.43
Birthweight (kg)	3.2 ± 0.1	3.2 ± 0.1	.57
LHR	0.97 ± 0.05	1.01 ± 0.04	.07
LHR (observed to expected lung to head ratio)	0.37 ± 0.02	0.34 ± 0.02	.59
Liver up position CDH (n, %)	37 (65%)	25 (66%)	>.99
Right sided CDH (n, %)	10 (18%)	2 (7%)	.32
PaO_2 at initiation (mm Hg)	65 ± 6	51 ± 3	.27
FiO_2 at initiation (%)	76 ± 4	78 ± 4	.71
P/F at initiation (mm Hg)	121 ± 16	82 ± 10	.45
A-a gradient at initiation (mm Hg)	397 ± 28	422 ± 30	.77
pH at initiation	7.18 ± 0.02	7.20 ± 0.02	.63
All right to left shunting on echo (n, %)	19 (37%) (n = 52)	8 (22%) (n = 37)	.16
Bowing ventricular septum (n, %)	50 (56%)	15 (43%)	.14
LV dysfunction (n, %)	14 (27%) (n = 52)	3 (8%)	.03

Lawrence et al, J Peds 2019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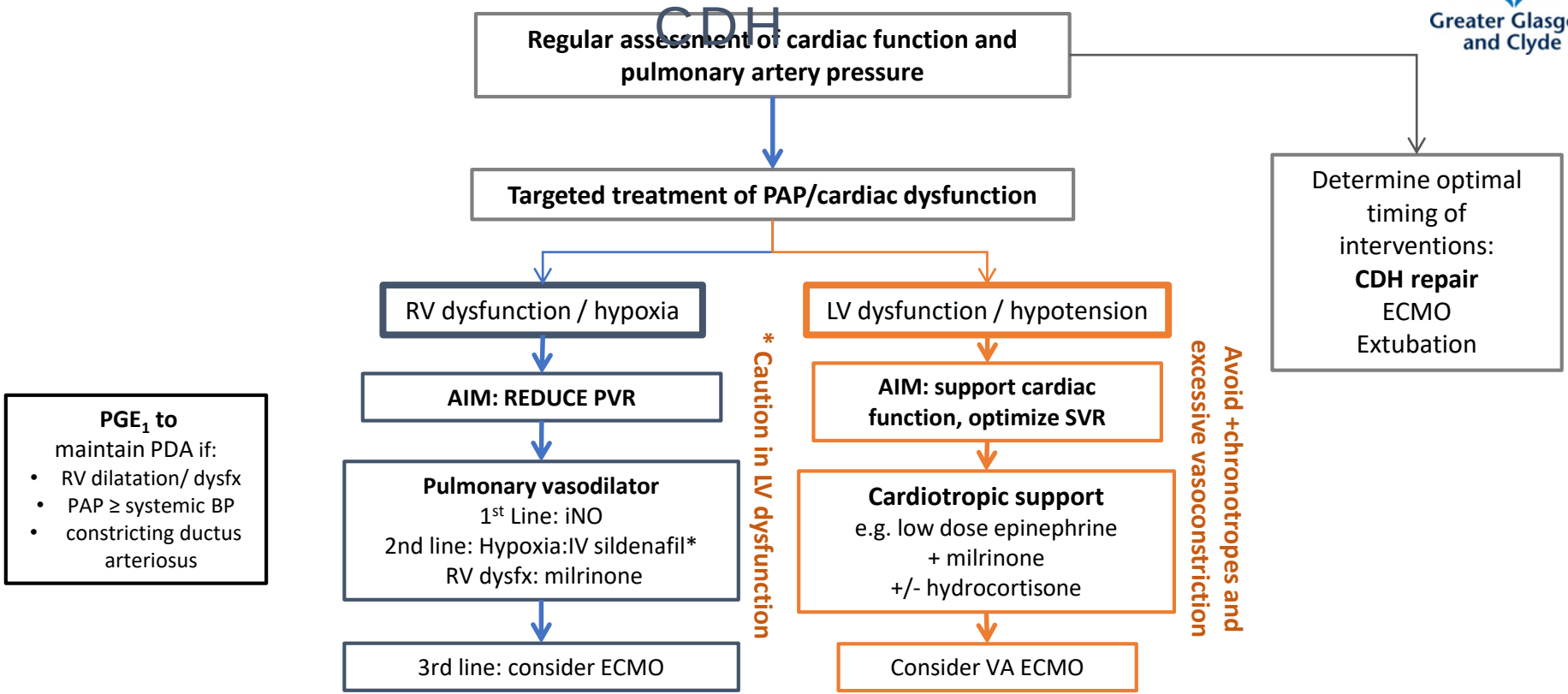
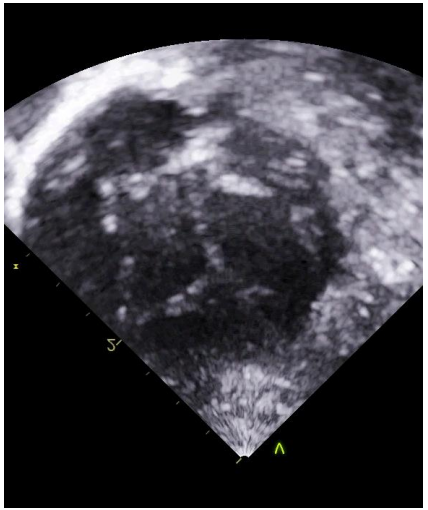
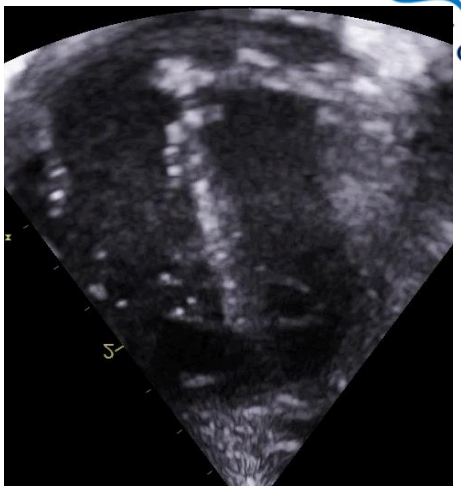
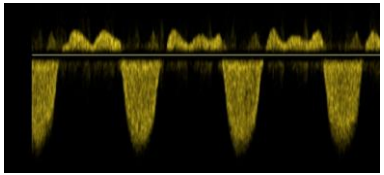
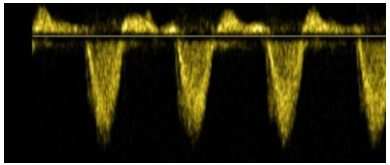
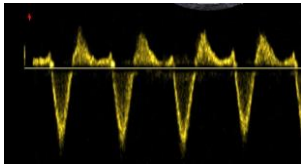
Fetal LV hypoplasia
↑ Pulmonary Vascular Resistance
↑pulmonary venous pressure
↑ afterload at birth
Right-to-left shunting (atria and PDA)
RIGHT VENTRICULAR DYSFUNCTION
Ventricular interdependence
LEFT VENTRICULAR DYSFUNCTION
Reduced cardiac output
Hypoxemia, acidosis, systemic hypotension

Targeted therapy of PH and cardiac function in CDH

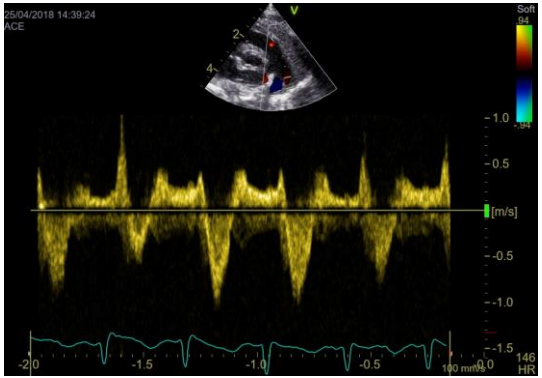


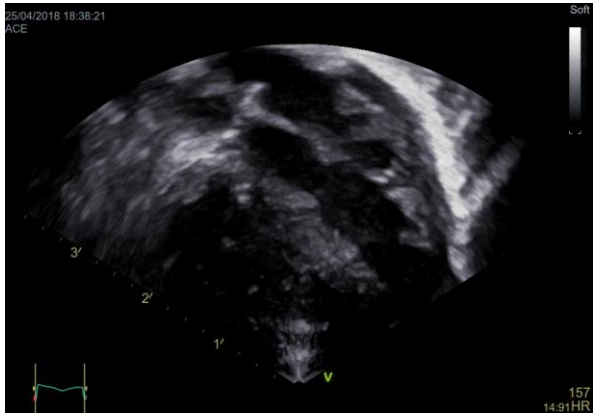
	Day 1	Day 3	Day 5
4 chamber view			
PDA flow			
STE Strain	RV GLS -12% LV GLS [illegible]%	RV GLS -12% [illegible]	RV GLS -16% [illegible]S -20%

Milrinone and epinephrine

IV sildenafil

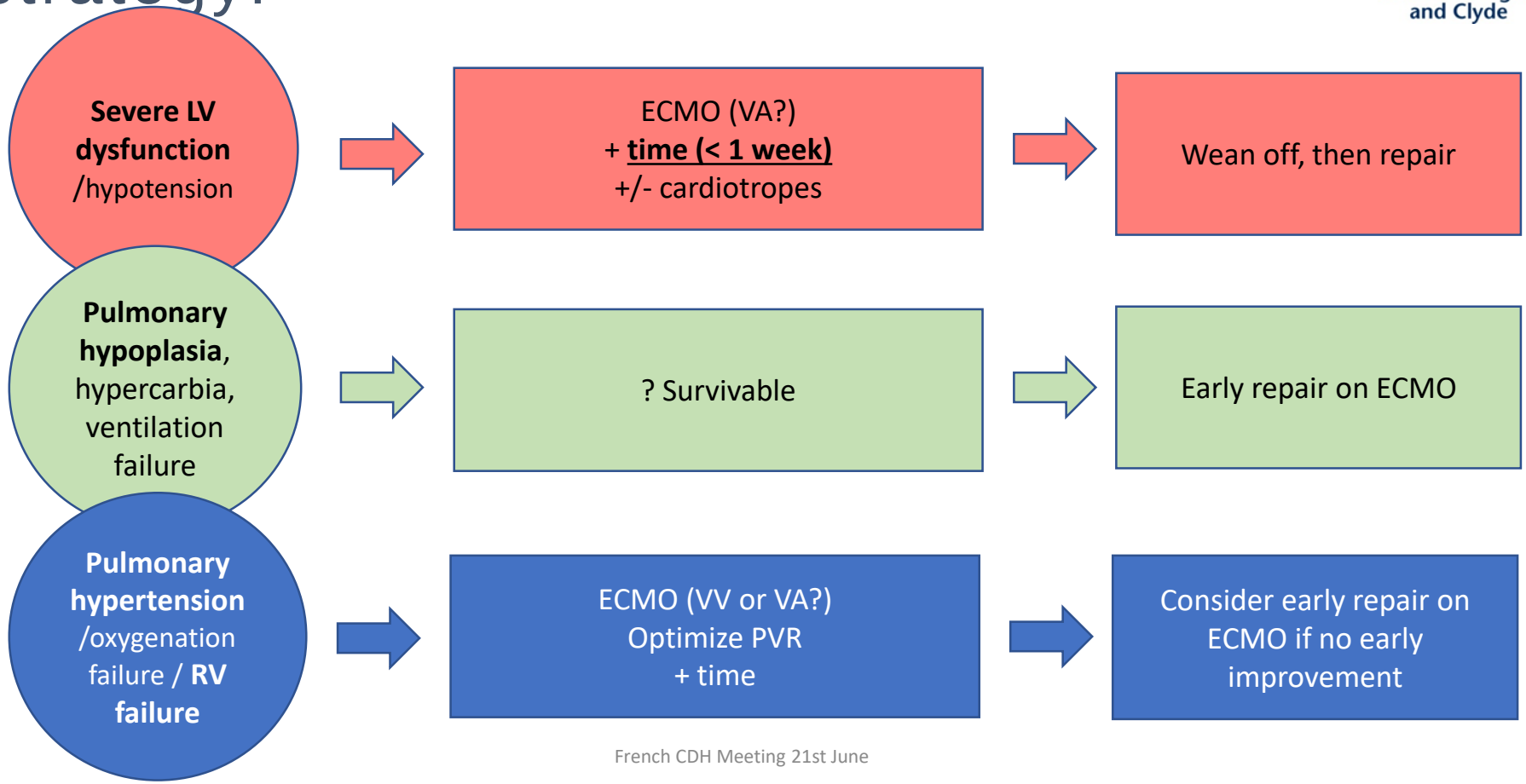
Cardiac function and timing of repair?





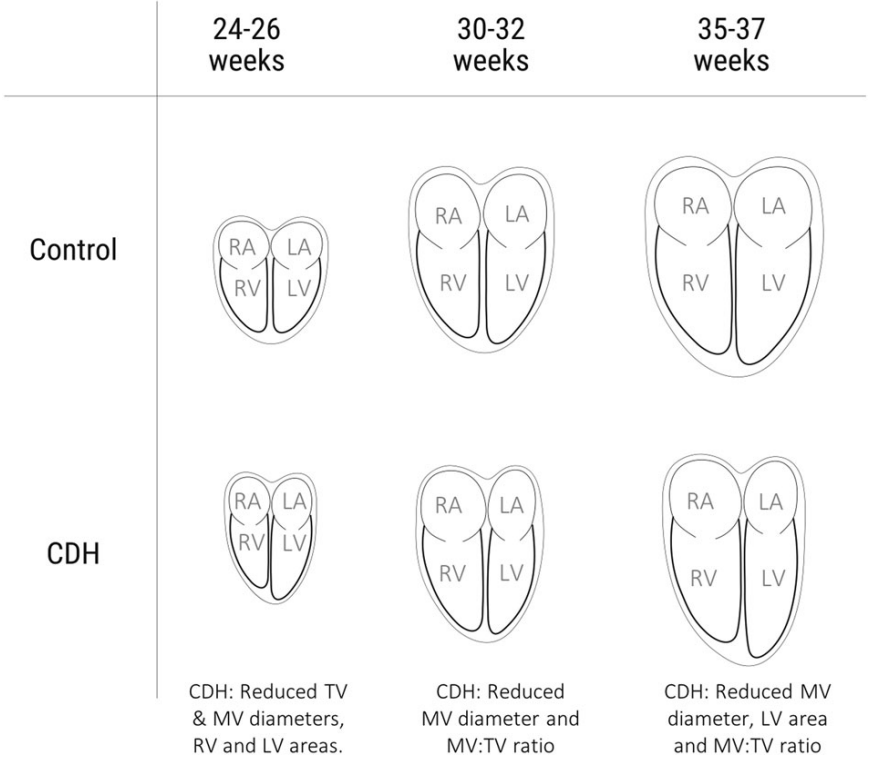
- 38/40. 3kg. L CDH. Day 1 of life
- Conventional ventilation: 22/5, FiO_2 0.35. Sats 96/97%. BP 45/32 (36)
- Mild/moderate LV dysfunction (Global Longitudinal Strain -14%)
- Cardiac function improved spontaneously by day 3 of life
- Primary repair, day 3. Stage “A” defect

CDH pathophysiology and ECMO strategy:



Fetal cardiac dimensions in congenital diaphragmatic hernia: relationship with gestational age and postnatal outcomes

Anna Claudia Massolo[1] · Anita Romiti[1] · Milena Viggiano[1] · Chiara Vassallo[1,2,3] · Marie Anne Ledingham[4] · Antonio Lanzone[5] · Leonardo Caforio[1] · Pietro Bagolan[1] · Neil Patel[5]



J Perinatol. 2021

Feasibility and safety of intact cord resuscitation in newborn infants with congenital diaphragmatic hernia (CDH)

Caroline Lefebvre[a,b], Thameur Rakza[a,c,e], Nathalie Weslinck[d], Pascal Vaast[c], Véronique Houfflin-debarge[c,d,e], Sébastien Mur[a], Laurent Storme[a,*], for the French CDH Study Group

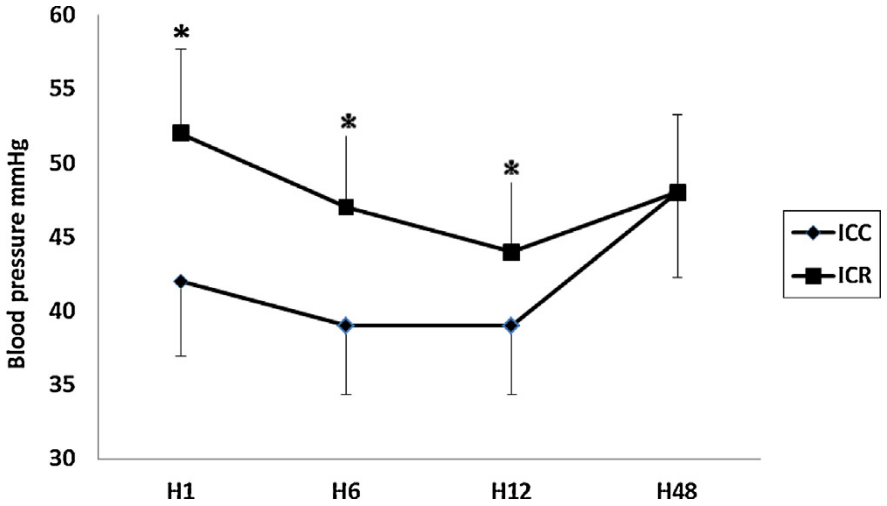
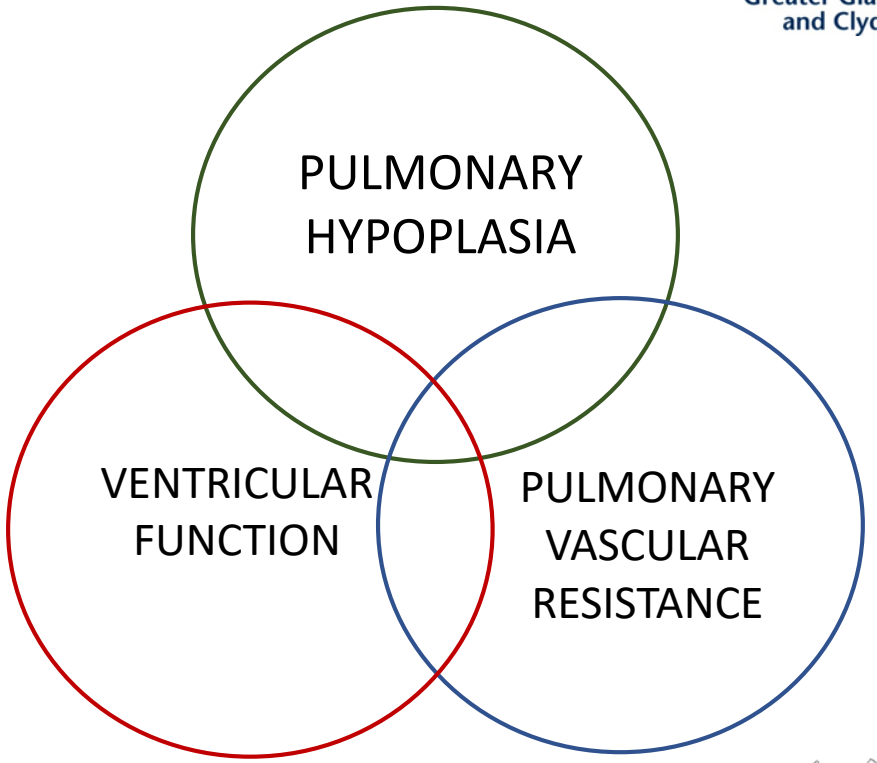


Fig. 2. Mean ± SD change in blood pressure (mmHg) after birth in immediate cord clamping (ICC) and intact cord resuscitation (ICR) groups. * $p < 0.05$ for comparison between groups.

C. Lefebvre et al. / Resuscitation 120 (2017) 20–25


PULMONARY HYPOPLASIA
VENTRICULAR FUNCTION
PULMONARY VASCULAR RESISTANCE

SAVE THE DATE!

CONGENITAL DIAPHRAGMATIC HERNIA INTERNATONAL SYMPOSIUM 2022

27-29th APRIL, 2022
GLASGOW, UK

NHS Greater Glasgow and Clyde

Thanks to

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Kevin Lally, Pam Lally, Matt Harting
CDH Study Group and Registry
CDH Euroconsortium
CDH UK

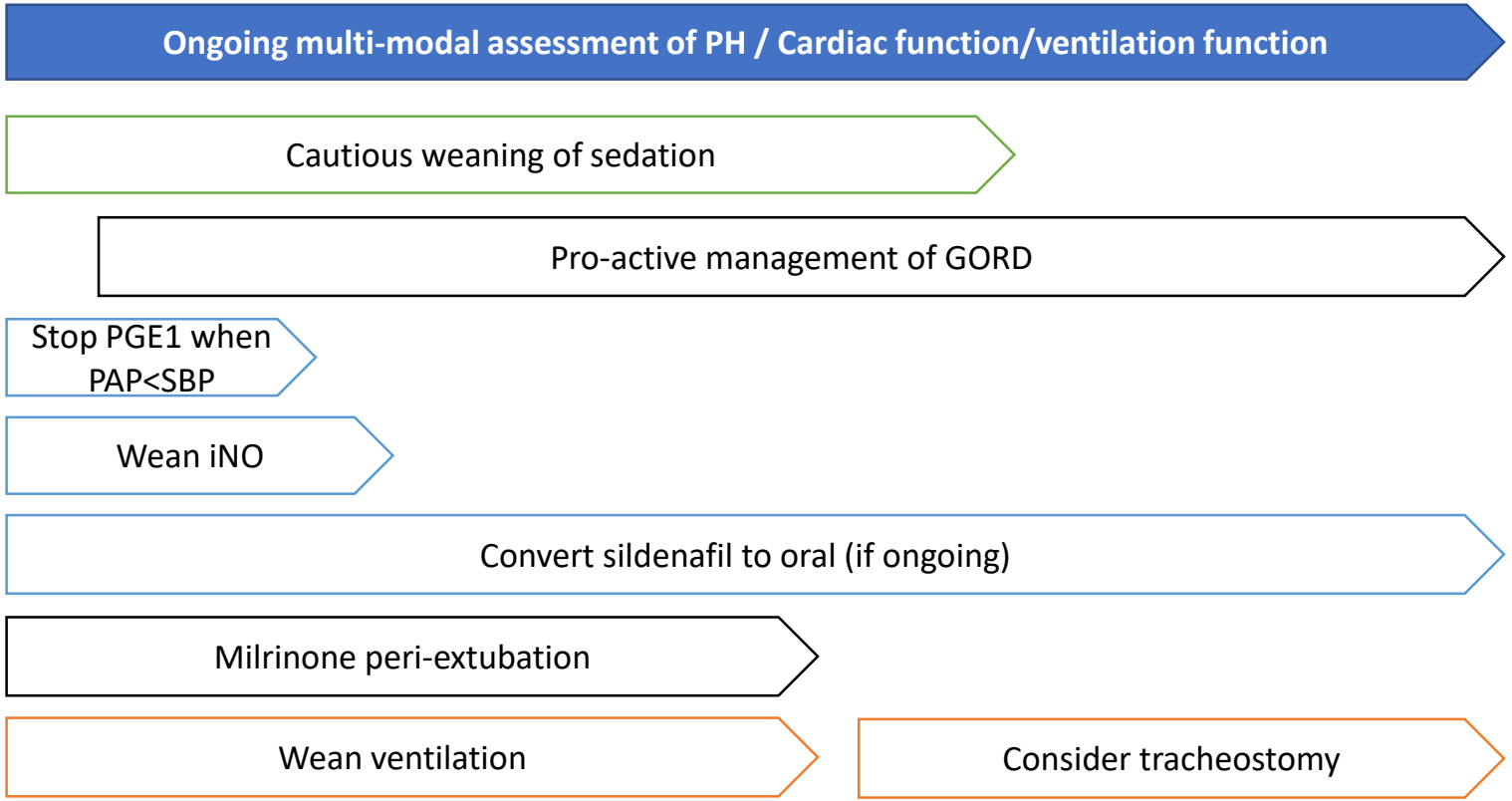


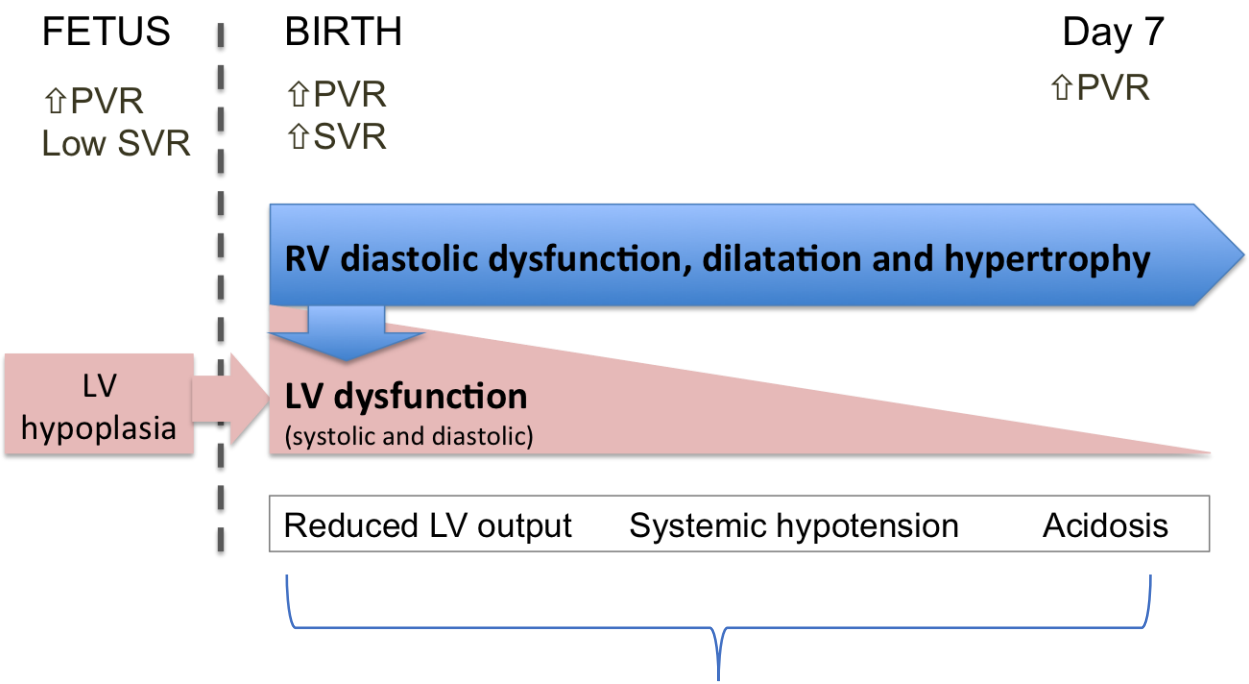






Post-operative management and weaning:





Associated with outcomes:

- Duration ventilation
- Length of stay
- Survival
- ECMO

N. Patel, F. Kipfmueller / Seminars in Pediatric Surgery 26 (2017)